What is claimed is:

1. A standardized prion preparation, comprising:

prions obtained from a plurality of animals; and

a carrier;

wherein the preparation is characterized by containing prions (a) which infect and cause disease in a known species of animal, (b) which are of a known strain, and (c) which are present in a known amount, and further wherein the carrier is of a known composition which is different from brain tissue of the mammal which the prions would infect in the animals natural state.

2. The preparation of claim 1, wherein the known amount is a known number of infectious units and known concentration of prions and wherein the known species is selected from the group consisting of human, cow and sheep.

3. The preparation of claim 1, wherein the preparation is a brain homogenate, the carrier is comprised of brain tissue from a mouse and wherein the prions infect and cause disease in a mammal selected from the group consisting of human, cow and sheep.

4. The preparation of claim 1, wherein substantially all prions present in the preparation are of a single strain and wherein the carrier is comprised of water..

5. The preparation of claim 4, wherein the strain has a polymorphism selected from the group consisting of: human M129, human V129, human E219, human K219 , sheep R171, sheep E171, sheep A136, sheep V136, a bovine 5 octarepeat polymorphism, and bovine 6 octarepeat polymorphism.

6. The preparation of claim 4, wherein the strain is human and has a pathogenic mutation selected from the group consisting of: a 2 octarepeat insert, a 4 octarepeat insert, a 5 octarepeat insert, a 6 octarepeat insert, a 7 octarepeat insert, an 8

octarepeat insert, a 9 octarepeat insert, P102L, P105L, A117V, D178N, V180I, F198S, E200K, V210I, D217R, M232A, and a stop codon at 145.

7. The preparation of claim 1, wherein the prions are of a plurality of different known strains and wherein the prions are obtained from 10 or more animals, and further wherein the prions are produced in a transgenic mouse having a genome comprising exogenous genetic material encoding at least a portion of a PrP protein.

8. The preparation of claim 1, wherein the prions are obtained from a mouse having an ablated endogenous PrP gene and a genome manipulated to express a high copy number of an exogenous PrP gene from a genetically diverse animal, and wherein the mouse spontaneously forms prions that normally infect the genetically diverse animal wherein the genetically diverse animal is selected from the groups consisting of a human, cow, sheep, dog, cat, goat, chicken or turkey.

9. The preparation of claim 1, wherein the prions are uniformly dispersed in the preparation and are produced in a transgenic mouse selected from the group consisting of: Tg(HuPrP), Tg(HuPrP)/Prnp$^{+/0}$ Tg(HuPrP)/Prnp$^{0/0}$, Tg(HuPrP^{CJD}), Tg(HuPrP^{CJD})/Prnp$^{+/0}$, Tg(HuPrP^{CJD})/Prnp$^{0/0}$, Tg(SHaPrP), Tg(SHaPrP$^{+/0}$)81/Prnp$^{+/0}$, Tg(SHaPrP)/Prnp$^{+/0}$, Tg(SHaPrP)/Prnp$^{0/0}$, Tg(ShePrP), Tg(ShePrP)/Prnp$^{+/0}$, Tg(ShePrP)/Prnp$^{0/0}$, Tg(BovPrP), Tg(BovPrP)/Prnp$^{+/0}$, and Tg(BovPrP)/Prnp$^{0/0}$.

10. The preparation of claim 1, produced in a transgenic mouse selected from the group consisting of: Tg(MHu2M), Tg(MHu2M)/Prnp$^{+/0}$, and Tg(MHu2M)/Prnp$^{0/0}$.

11. A prion protein standard composition comprising:

isolated exogenous prions from a plurality of transgenic mice genetically manipulated to allow infection by prions that normally only infect a genetically diverse animal, said mice being infected with prions of the genetically diverse animal; and

brain homogenate from the genetically diverse animal;

wherein the standard has properties sufficiently established to serve as reference control for prion measurement protocols.

12. The composition of claim 11, where prion measurement protocols are selected from the group consisting of: calibration of an apparatus, calibration of an assay, determination of specificity of an assay, determination of sensitivity of an assay, and determination of quality of an assay reagent.

13. The composition of claim 11, wherein the properties are selected from the group consisting of: a known endogenous host prion protein concentration, a known number of infectious units, a known exogenous host prion protein concentration, a known endogenous prion protein from the genetically diverse animal, a known genetic homogeneity, a known sensitivity for an antibody, and a known background protein concentration.

14. The composition of claim 11, wherein the concentration of the prions in the standard is augmented by the addition of isolated prion protein and wherein the transgenic host mammal is a mouse and the genetically diverse animal is selected from the group consisting of: human, cow, sheep, dog, cat, goat, chicken or turkey.

15. The composition of claim 11, further comprising:

isolated exogenous prions from a second plurality of transgenic mice genetically manipulated to allow infection by prions that normally only infect a second, different genetically diverse animal, said mice infected with prions that infect the second genetically diverse animal wherein the prions from the first plurality of mice are separate from the prions from the second plurality of mice and further wherein the prions are uniformly dispersed in the preparation.

16. The composition of claim 15, further comprising brain homogenate from the second genetically diverse animal.

17. A method for preparing a prion protein standard comprising the steps of:

providing a plurality transgenic host animals having an ablated endogenous PrP gene, said host animal genetically manipulated to allow infection by prions that normally only infect a genetically diverse animal;

inoculating the host animals with a composition comprising a known amount and strain of prions which infect the genetically diverse animal;

incubating the host animals until they exhibit sufficient symptoms of prion infection;

harvesting the brain tissue from the host animals exhibiting sufficient symptoms of prion infection; and

homogenizing the harvested brain tissue from the host animals;

wherein the homogenized prion preparation is characterized by having a known amount of prions.

18. The method of claim 17, wherein the known amount of prions of the homogenized prion preparation is a known number of infectious units, and wherein the number of infectious units is between 0.1 and about 100.

19. The method of claim 18, wherein the number of infectious units is between about 1 and about 10 and wherein the transgenic host animals are mice and the genetically diverse animal is selected from the group consisting of: a human, cow, sheep, dog, cat, goat, chicken or turkey.

20. A method of calibration of a prion protein assay, comprising the steps of:

providing a prion protein standard;

determining a true value of prion protein concentration in the prion standard;

subjecting a portion of the preparation to prion protein assays to determine an assay value for the standard;

determining a correction value for the assay based on the true value; and

adjusting the assay value to reflect the true level of prion protein in the standard;

wherein the assay is calibrated by adjusting the assay value to reflect the true value of prion protein concentration in a sample.

21. The method of claim 20, wherein the assay is calibrated using a plurality of standards with different prion protein concentrations.

22. The method of claim 20, further comprising the steps of:

subjecting a portion of the preparation to a second prion protein assay to determine a second assay value for the standard;

determining a correction value for the second assay based on the true value; and

adjusting the second assay to reflect the true level of prion protein in the standard; and

comparing the adjusted levels of prion protein in each assay;

wherein the assays are calibrated with respect to one another by adjusting the assay values detected by each assay to reflect the true values of prion protein concentration.

23. A prion protein standard kit, comprising a plurality of protein preparations, each preparation characterized by containing prions (a) which infect and cause disease in a known species of mammal, (b) which are of a known strain, (c) which are present in a known amount, (d) which are obtained from a plurality of animals.

24. The kit of claim 23, wherein each preparation contains a different amount of prions characterized by the ability to infect and cause disease in the same species.

25. A kit of claim 23, wherein each preparation contains a different amount of a single strain of prion.

26. The kit of claim 23, wherein each preparation has a same known amount of a different prion strain.

27. The kit of claim 26, wherein each prion strain is characterized by the ability to infect and cause disease in a different species.

28. The kit of claim 26, wherein each preparation has one infectious unit of prions.